[BRIEF REPORT]

Telangiectasia Macularis Eruptiva Perstans Presenting as Island Sparing

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ABSTRACT

Mastocytosis is characterized by the proliferation and accumulation of mast cells within organs and most commonly the skin; localization accounting for the frequent presentation of skin lesions in affected individuals. The authors detail a case report involving a patient with telangiectasia macularis eruptive perstans, a rare cutaneous form of mastocytosis, accompanied by an unusual clinical finding of island sparing. (*J Clin Aesthet Dermatol.* 2013;6(4):41–42.)

Island sparing is a clinical feature of various dermatoses, including cutaneous T-cell lymphoma (CTCL), pityriasis rubra pilaris, and psoriasis, in which a distinct area of uninvolved skin exists within a larger lesion.[1–2] In this case, the authors present an adult male diagnosed with telangiectasia macularis eruptiva perstans (TMEP), a rare representation of cutaneous mastocytosis, accompanied by an unusual clinical presentation of island sparing.

Mastocytosis is a disorder marked by the proliferation and accumulation of mast cells within various organs. Mastocytosis presents in two forms—cutaneous and systemic.[3] Mast cells most commonly localize to the skin, accounting for the frequent presentation of skin lesions in patients affected by either form. Cutaneous mastocytosis typically develops early in life. Skin lesions observed in pediatric patients tend to involute by adolescence, while lesions in adults are often persistent and at higher risk of being associated with systemic disease.[4] Symptoms of systemic involvement include flushing, dyspnea, abdominal pain, diarrhea, hypotension, syncope, splenomegaly, and bone pain.[3–5]

Cutaneous mastocytosis can be divided into the following variants: urticaria pigmentosa, mastocytoma, diffuse and erythrodermic cutaneous mastocytosis, and TMEP.[3] TMEP is a rare form of cutaneous mastocytosis, occurring predominantly in adults. Lesions are diffuse and present as red telangiectatic macules accompanied by hyperpigmentation, typically on the trunk and extremities.[3] The urtication of lesions after firm stroking, a change known as Darier's sign, is often negative in patients with TMEP due to lesions being paucicellular.[4–5] Systemic involvement is uncommon, affecting only a small percentage of patients diagnosed with TMEP; however, all adult-onset cutaneous mastocytosis patients should be tested.[5] Laboratory testing includes serum tryptase and 24-hour measurement of urinary n-methylhistamine and prostaglandin D2 metabolites.[4]

A 60–year-old Caucasian man with a history of dermographism presented with an asymptomatic macular rash he developed over the past 10 years. Physical examination revealed multiple, red-tan, blanchable, 2 to 3mm macules present on the patient's entire trunk. Remarkable findings include macules taking on a confluent nature with the exception of an unusual finding of a single area of island sparing on the patient's right flank (Figure 1). Lesions clinically manifested as telangiectatic macules with a generalized erythematous background characteristic of TMEP (Figure 2) and were determined to cover 36 percent of the patient's body surface area using standard formula. The patient denied pruritus, and Darier's sign was negative. The finding of island sparing

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validated a differential diagnosis of CTCL.

A 3mm punch biopsy of a right flank lesion was consistent with TMEP. Histopathological examination with application of Giemsa and Leder stains revealed increased perivascular and interstitial mast cells surrounding dilated telangiectatic blood vessels typical of TMEP (Figure 3). Hematoxylin and eosin stain further revealed dilated telangiectatic blood vessels with a mild perivascular infiltrate of mononuclear mast cells. Histological features of CTCL were completely absent.

Although symptoms of systemic involvement other than occasional diarrhea were not reported, the patient was referred for additional laboratory testing. Serum tryptase, bone marrow aspirate/biopsy, and complete blood count were negative for systemic mastocytosis.

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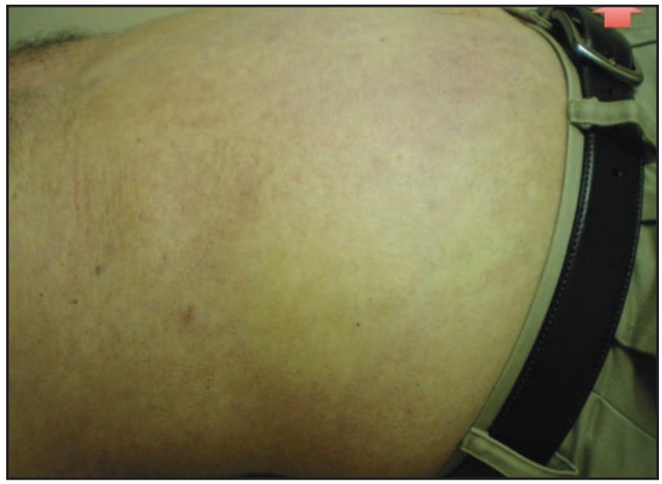

Figure 1. Unusual physical manifestation of TMEP in which the patient's entire trunk is involved with the exception of island sparing on the right flank

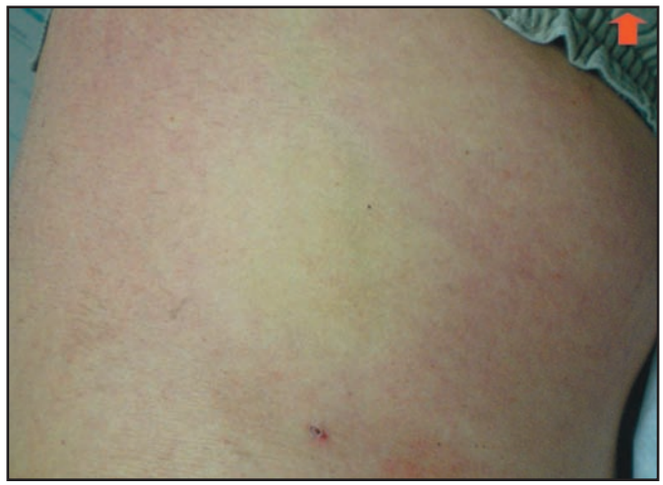

Figure 2. Right flank displaying confluent telangiectatic macules surrounding round area of island sparing. Biopsy site can be seen

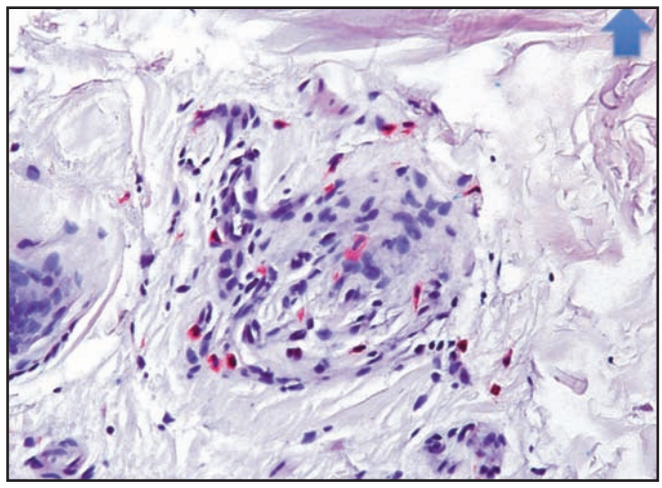

Figure 3. A Leder stain highlights a mild increase in the number of mast cells, some of which are rounded, others are spindled. A few of the vascular spaces are telangiectatic. Features of T-cell lymphoma are lacking